The Lemierre syndrome - a neuroophthalmological approach

Vadasz, Vinzenz A C ; Gerth-Kahlert, Christina

Abstract: Case report of a twenty-two year old woman with double vision after tonsillitis, caused through multiples thrombosis by an infection with fusobacterium necrophorum known as the Lemierre-Syndrome. Fig. 1: Ocular motility at ICU (lying position) Fig. 2: white arrows show thrombosis of the right ophthalmic vein Fig. 3: white arrows show retropharyngeal mass Fig. 4: White arrows show thrombosis of the inner jugular veins Fig. 5: Brain-MRA shows fusiform aneurysm of the right internal carotic artery in the cavernous segment Fig. 6: Demonstrates palsy of the left abducens nerve Fig. 7: Demonstrates normal eye position and movement Fig. 8: Prof. Lemierre

The Lemierre Syndrome
A Neuroophthalmological Approach

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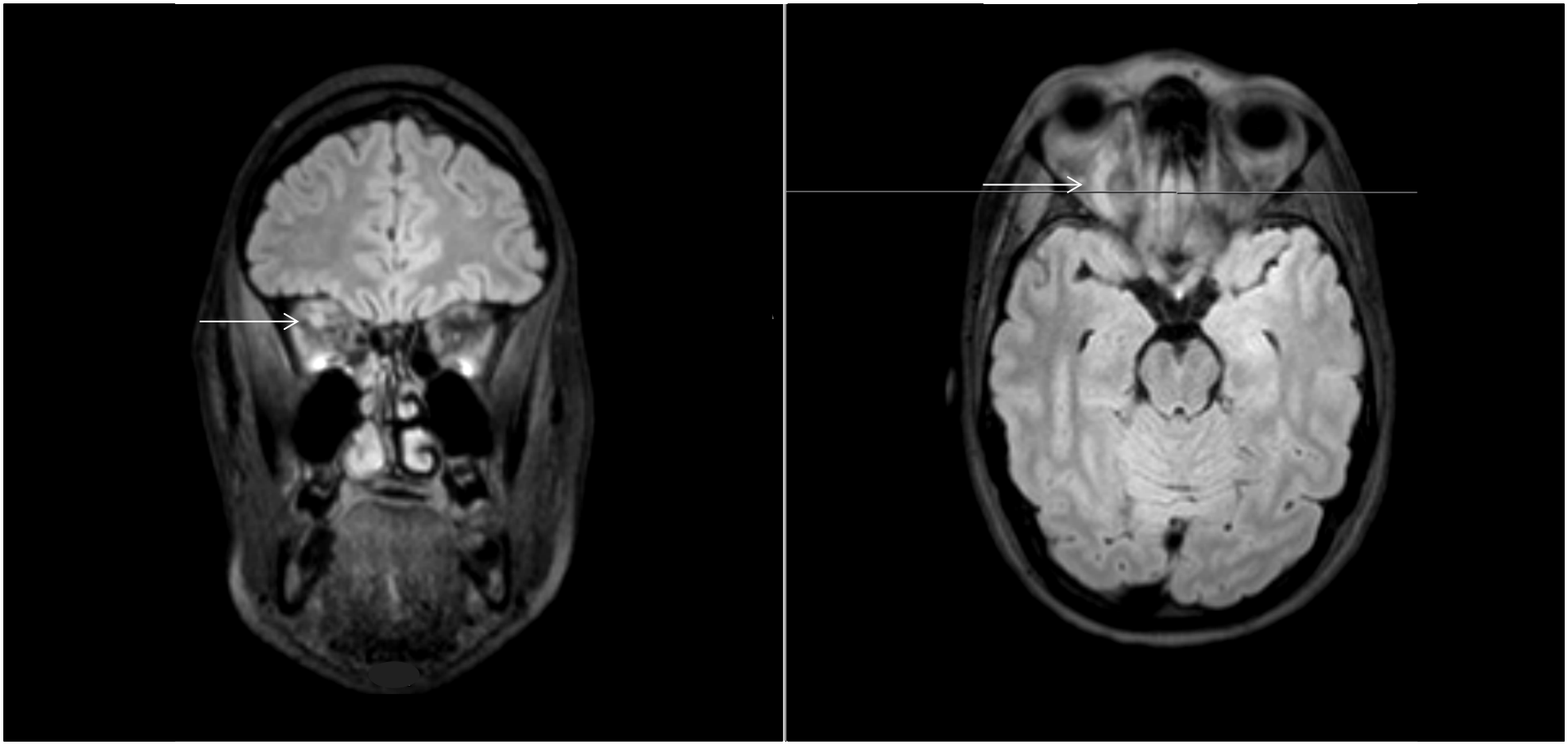
Case Report

- Twenty-two year old female patient
- Diagnosed with acute tonsillitis 6 days ago
- Symptoms: diplopia, photophobia
- Signs:
  - full visual acuity
  - OU: VI nerve palsy
  - OD: partial III nerve palsy
  - OD exophthalmus 2.5mm
  - incomplete trismus
  - discrete signs of meningism
  - fever

Fig. 1: Ocular motility at ICU (lying position)

Head MRI (flair) 3 days after symptom onset

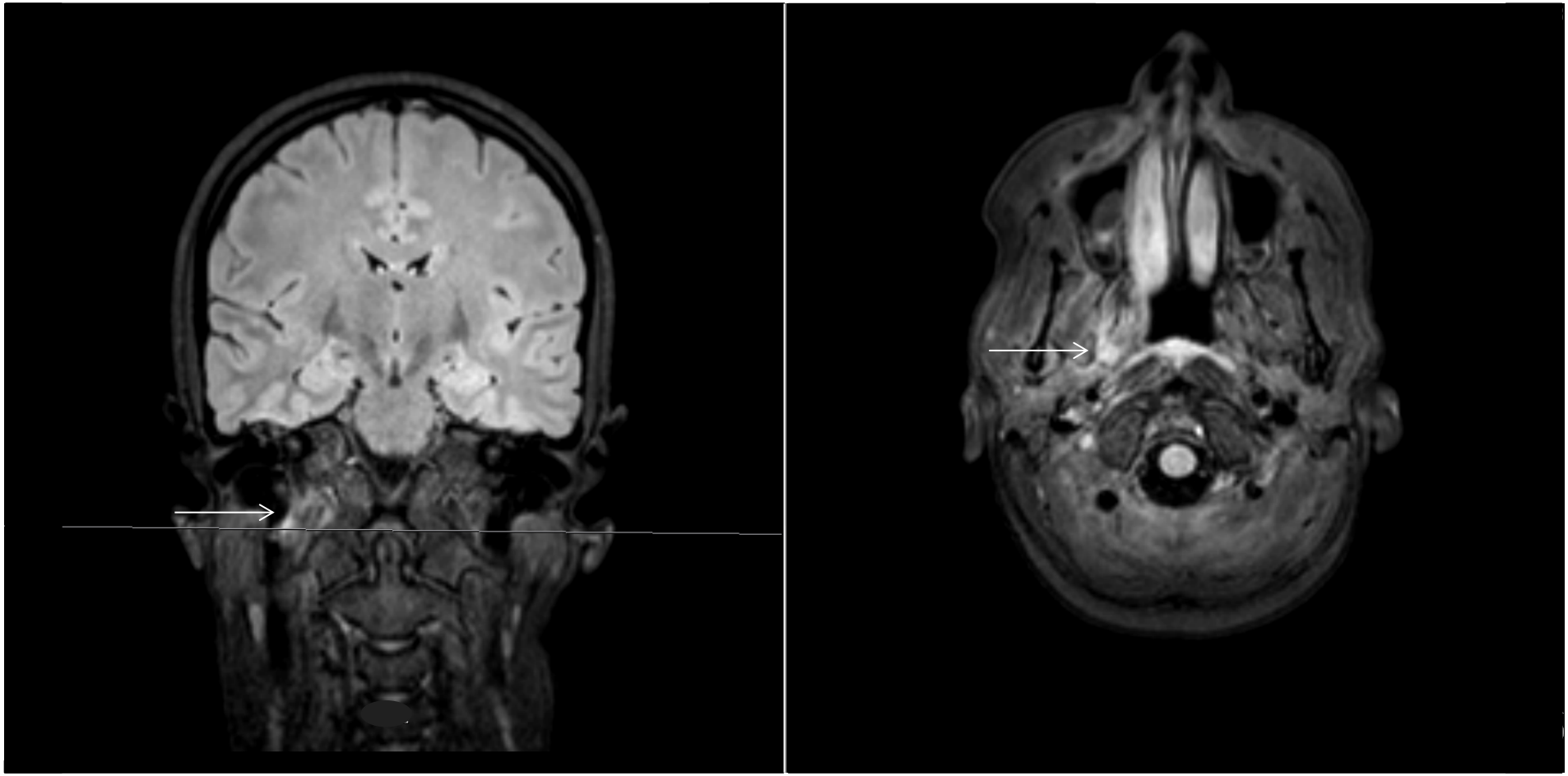


Coronal plane

Transverse plane

Fig. 2: white arrows show thrombosis of the right ophthalmic vein

Head MRI (flair) 3 days after symptom onset

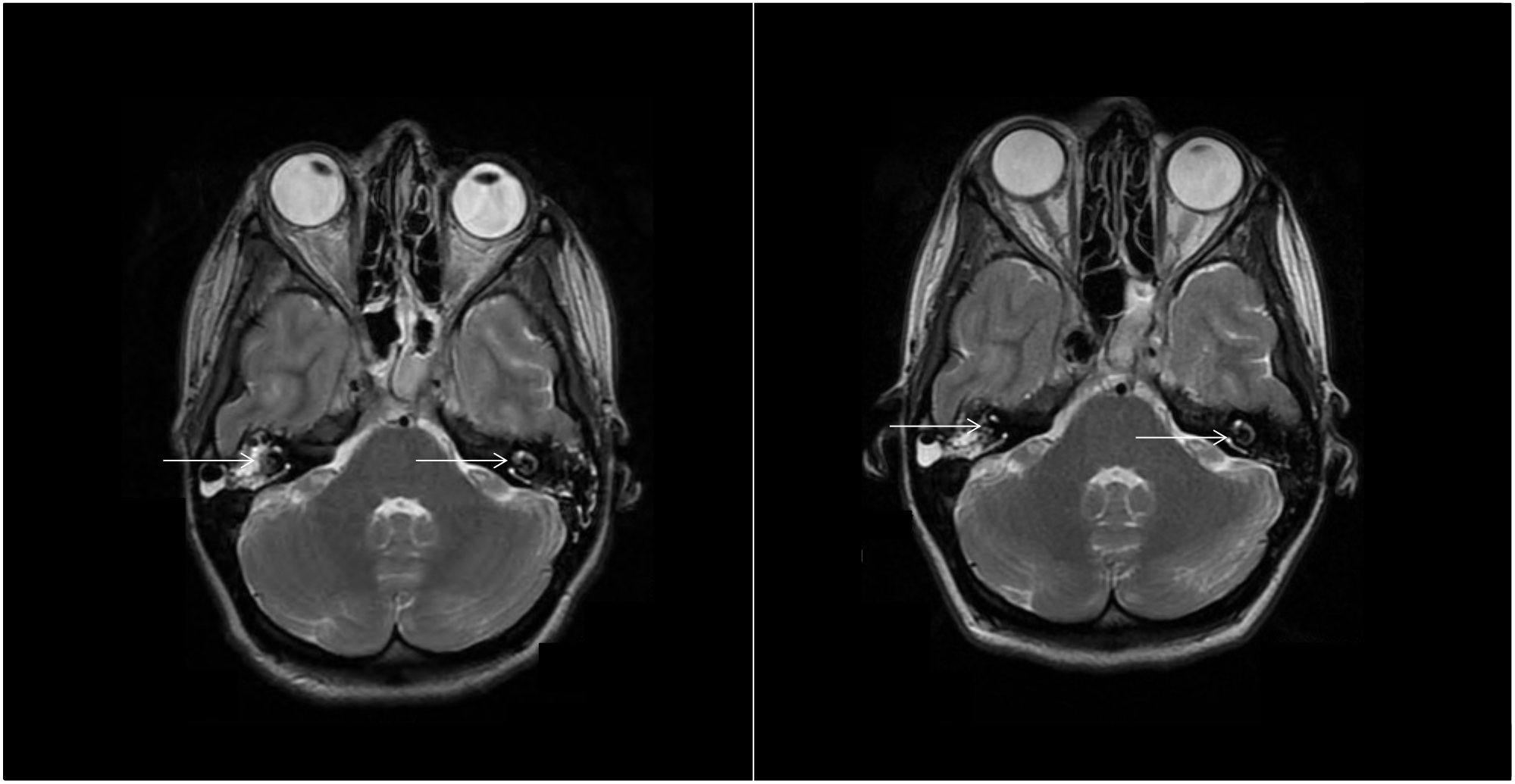


Coronal plane

Transverse plane

Fig. 3: white arrows show retropharyngeal mass

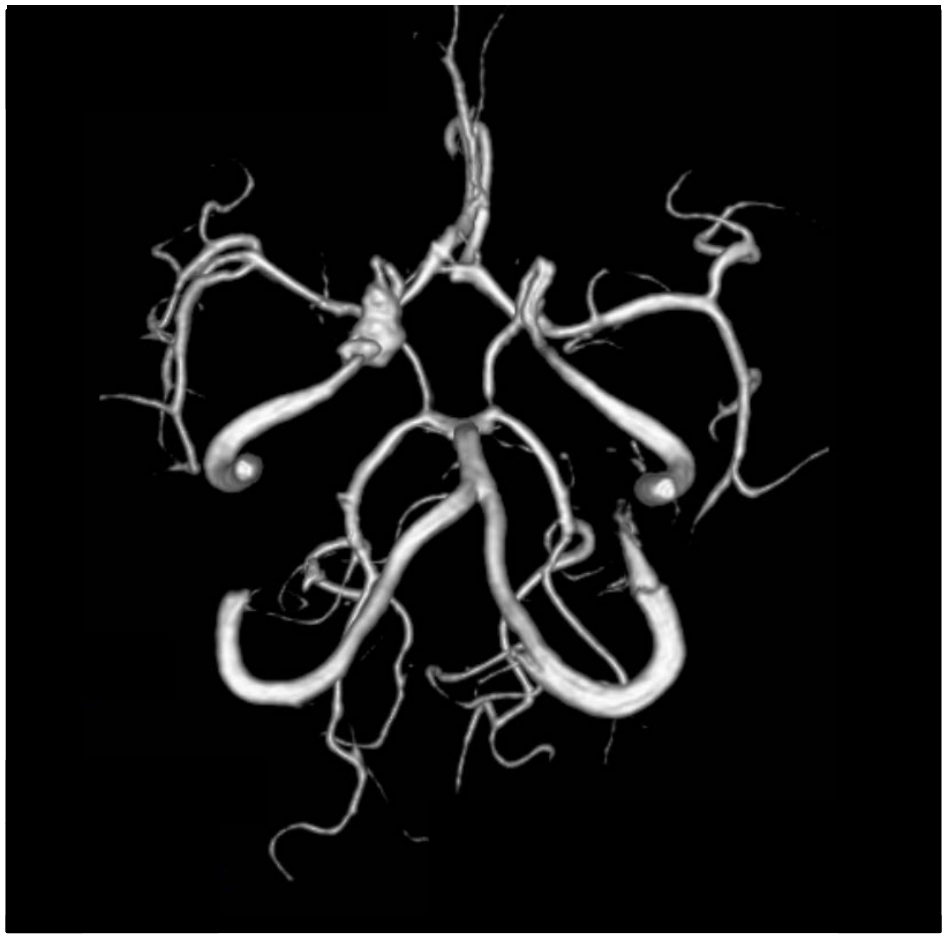
Head MRI (flair) 10 days after symptom onset



Transverse plane

Fig. 4: White arrows show thrombosis of the inner jugular veins

Head MRA 14 days after symptom onset



Bottom view

Fig. 5: Brain-MRA shows fusiform aneurysm of the right internal carotid artery in the cavernous segment

Diagnostics

Laboratory:

Leucocytosis: 16.000/µl (15.000/µl neutrophils, 45% left shift)

C-reactive protein (CRP): 271 g/l

Thrombocytopenia (21,000/µl)

International normalized ratio (INR): 1.27

Blood cultures:

Fusobacterium necrophorum was found in blood cultures

Diagnoses

Lemierre Syndrome with:

1. **Septic thrombosis:**

- Bilateral cavernous sinus

- Right superior ophthalmic vein

- Bilateral sphenoparietal sinus

- Bilateral internal jugular vein

- Right subclavian vein with thrombus

- Right retromandibular vein and basis of the skull

2. Bilateral **septic pulmonary embolism**

3. **Brain abscess,** right temporo-basal area

4. **Basal meningitis**

5. Vascular change of the left carotic artery

   fusiform aneurysm of the right internal carotic artery in the cavernous segment

6. **Sphenoidal sinusitis**

- transnasal left sphenoidectomy

Hospital course

5 months after symptom onset

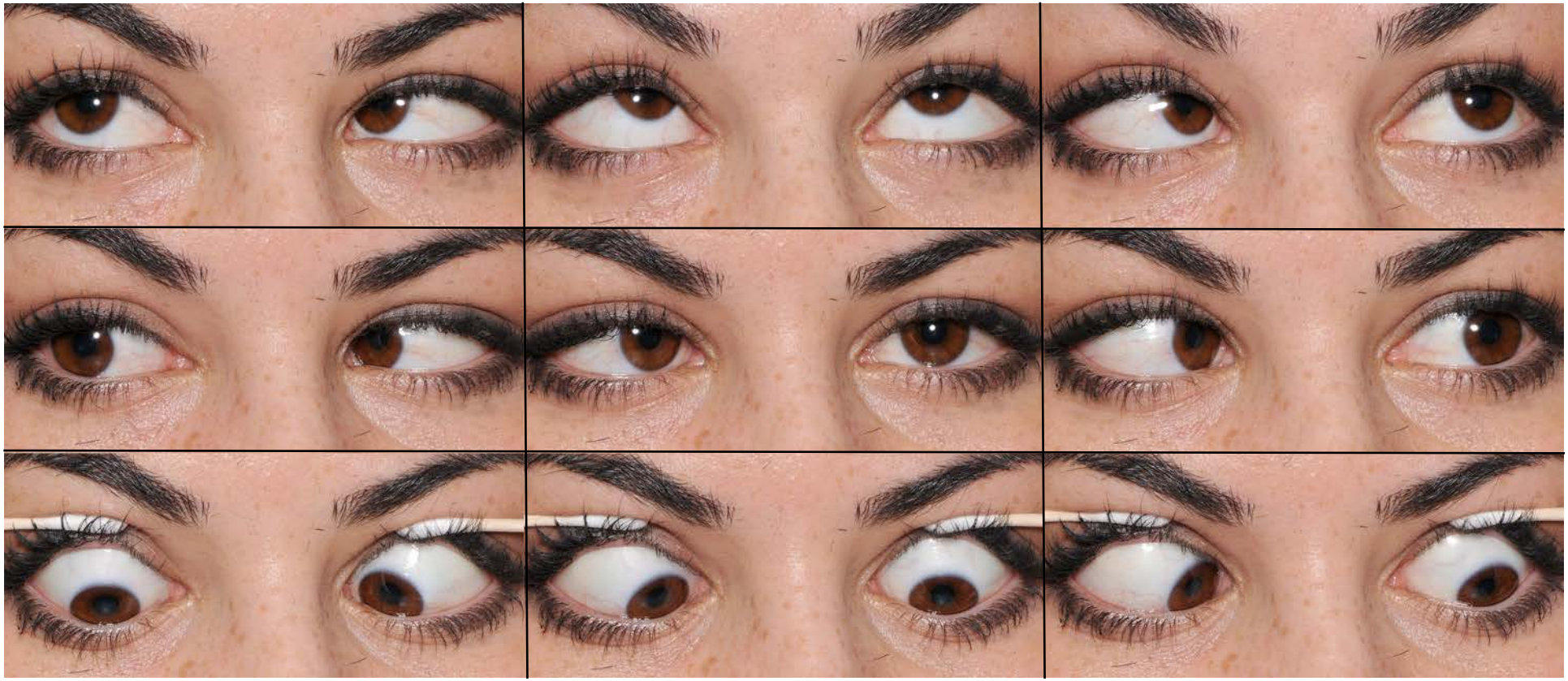


Fig. 6: Demonstrates palsy of the left abducens nerve

26 months after symptom onset, 12 months after strabismus surgery

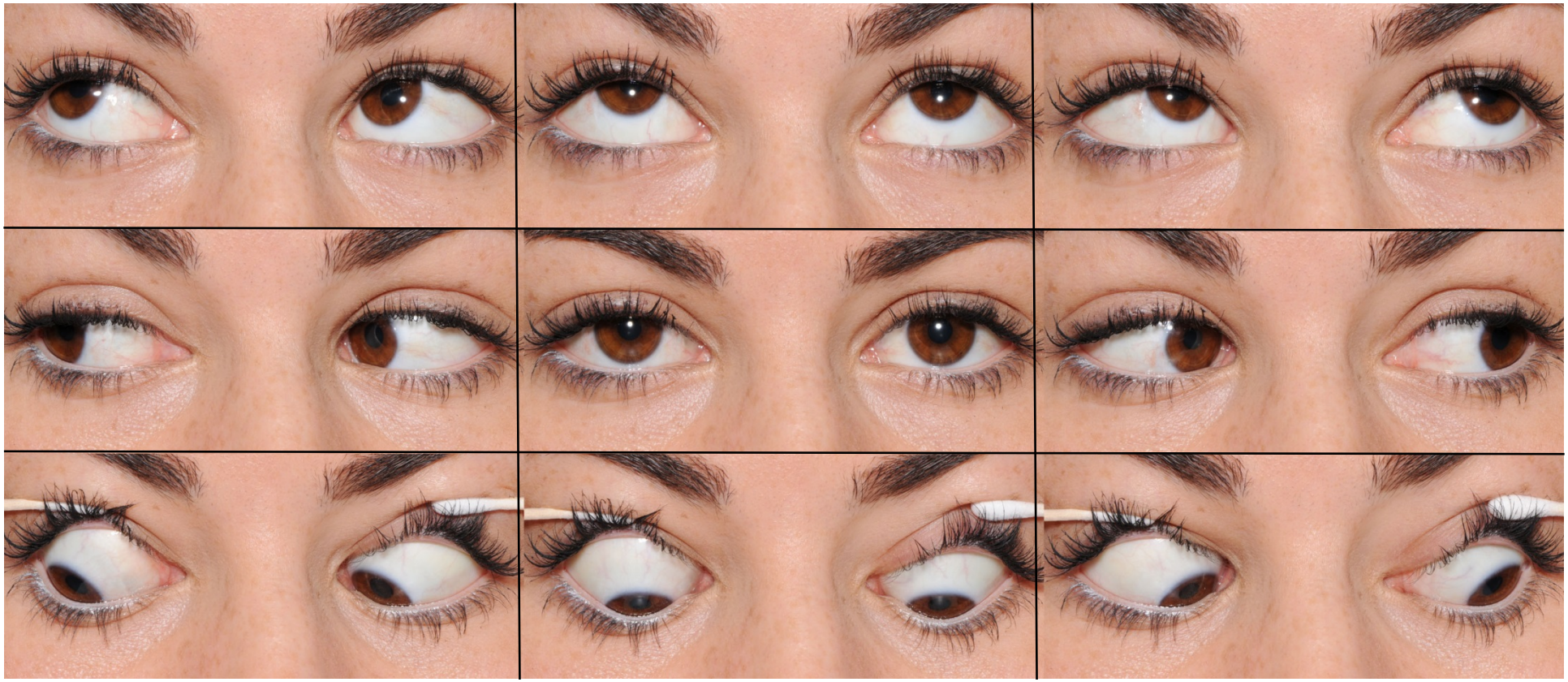


Fig. 7: Demonstrates normal eye position and movement

Lemierre Syndrome: Background

Lemierre Syndrome

(Postanginal septicemia)

- Acute oropharyngeal infection with secondary septic thrombophlebitis of the internal jugular vein (IJV) and frequent metastatic infections. [1]
- The common etiologic agent is *Fusobacterium necrophorum*[1]

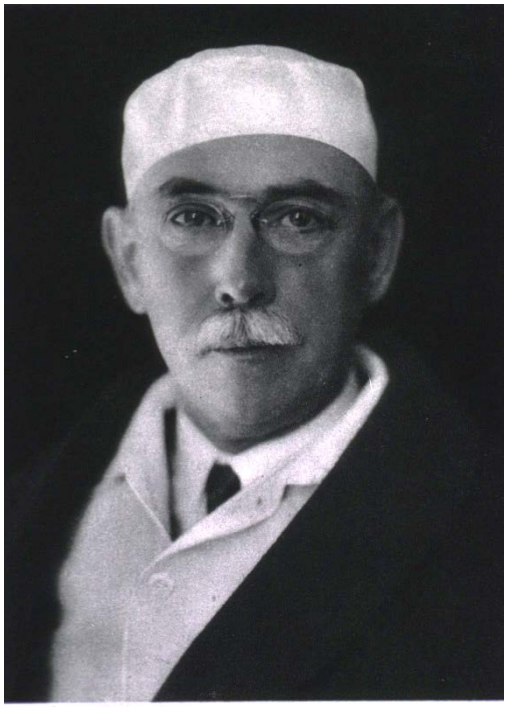


Fig. 8: Prof. Lemierre[2]

References:

1) Medicine (Baltimore). 2002 Nov;81(6):458-65. **The evolution of Lemierre syndrome: report of 2 cases and review of the literature.** Chirinos JA

2) Foto: http://ihm.nlm.nih.gov/images/B17177

Fusobacterium necrophorum

- Normal flora of the oral cavity, the female genital tract and the gastrointestinal tract[1]
- Strictly anaerobe, non-motile, non-spore-forming, Gram-negative rod[1]
- Was found in 61-100% of the cases (blood)[2]
- Ability to cause severe disease as a primary pathogenic agent[1]

References:
1) Medicine (Baltimore). 2002 Nov;81(6):458-65. **The evolution of Lemierre syndrome: report of 2 cases and review of the literature.** Chirinos JA
2) Postgrad Med J 2004 Jun;80(944):328-34. **Lemierre's syndrome: more than a historical curiosa.** Riordan T

Pathogenesis

- Primary infection
  - Palatine tonsils/peritonsillar tissue (87.1%) with sore throat
  - Fever (82.5%), not necessarily at the time of initial presentation
  - < 1 week between onset of oropharyngeal infection and second stage

Reference:
Medicine (Baltimore). 2002 Nov;81(6):458-65. **The evolution of Lemierre syndrome: report of 2 cases and review of the literature.** Chirinos JA

Pathogenesis

- Metastatic complications
  - Bacteremia with hematogenous spread
  - most commonly to the lungs (79.8%) and joints (16.5%)
  - Chest X-ray with pleural effusion (43.1%)
  - Normal chest X-ray (19.2%)

Reference:
Medicine (Baltimore). 2002 Nov;81(6):458-65. **The evolution of Lemierre syndrome: report of 2 cases and review of the literature.** Chirinos JA

Pathogenesis

- Second stage
  - Infection spreads from peritonsillar tissue to the adjacent lateral pharyngeal space via lymphatic vessels
  - Thrombophlebitis of the IJV, severe sepsis with metastatic infections
  - Swollen and/or tender neck complaints (52.2%)
  - No significant neck findings (47.7%)
  - Horner syndrome (sympathetic trunk involvement), carotid artery rupture

Reference:
Medicine (Baltimore). 2002 Nov;81(6):458-65. **The evolution of Lemierre syndrome: report of 2 cases and review of the literature.** Chirinos JA

Reported neuroophthalmic complications

- Cranial nerves III[1], IV[2], VI[3], IX – XII[3]
- Horner syndrome[3]
- Chemosis[4], ptosis[4], septic orbital vein thrombosis[4,5,6], proptosis[5], orbital cellulitis[6]
- Endophthalmitis[7]

References:

1) Nervenarzt. 2003 Dec;74(12):1118-21. **[Bacterial meningitis as a complication of Fusobacterium necroforum infection in adults].** Spitzer C
2) J AAPOS. 2009 Feb;13(1):107-8. doi: 10.1016/j.jaapos.2008.08.004. Epub 2008 Oct 18. **Lemierre's syndrome with fourth nerve palsy.** Lee S
3) Laryngoscope. 2007 Sep;117(9):1605-10. **Lemierre syndrome: two cases and a review.** Syed MI
4) Neurology. 2013 Sep 24;81(13):1179-80. **Lemierre syndrome: more than "the forgotten disease".** Morelli N
5) Ophthal Plast Reconstr Surg. 2011 May-Jun;27(3):e67-8. **Orbital dissemination of Lemierre syndrome from gram-positive septic emboli.** Kahn JB
6) Auris Nasus Larynx. 2013 Oct;40(5):493-6. **Blindness caused by septic superior ophthalmic vein thrombosis in a Lemierre Syndrome variant.** Akiyama K
7) Eye (Lond). 2004 Aug;18(8):860-2. **Endogenous endophthalmitis secondary to Lemierre's syndrome.** Ahad MA

Clues to Diagnosis

- Recurrence of severe pyrexial attacks several days after onset of a sore throat (tonsillary abscess) with initial rigor, or - more distinctly - the occurrence of pulmonary infarctions and arthritic manifestations.
- The first clue from blood cultures: Fusobacterium necrophorum (69.7%)

Reference:
Medicine (Baltimore). 2002 Nov;81(6):458-65. **The evolution of Lemierre syndrome: report of 2 cases and review of the literature.** Chirinos JA

Management

- Blood cultures
- CT as primary diagnostic method
- Ultrasound for follow-up
- MRI for infections arising from the mastoid (intracerebral vein thrombosis)
- Metronidazole or Clindamycin for 3-6 weeks
- Surgical ligation or excision of the IJV in patients with uncontrolled sepsis and ongoing septic emboli

Reference:
Medicine (Baltimore). 2002 Nov;81(6):458-65. **The evolution of Lemierre syndrome: report of 2 cases and review of the literature.** Chirinos JA